

PARENT FACT SHEET

DISORDER

Isovaleric Acidemia (IVA)

CAUSE

IVA occurs when an enzyme called “isovaleryl-CoA dehydrogenase” is either missing or not working properly. This enzyme’s job is to help break down a harmful substance called isovaleric acid. It is made in the body when the amino acid, leucine, is broken down. When a child with IVA eats food containing leucine, isovaleric acid builds up in the blood and causes problems. Leucine is found in all foods that contain protein.

IF NOT TREATED

Infants with IVA seem healthy at birth. Often, the first symptoms start between one day and two weeks of age. IVA causes episodes of illness called metabolic crises. If not treated, many babies die during their first metabolic crisis. In those who survive, repeated episodes of metabolic crisis can cause brain damage. This can result in life-long learning problems or mental delays.

TREATMENT OPTIONS

Your child will need to be under the care of a metabolic specialist and dietician. Treatment is usually needed throughout life.

- The IVA infant will require a special formula, which will be prescribed by the metabolic specialist and the dietician.
- As your child gets older, the dietician will develop a special food plan, which may include special medical foods.
- Glycine is an amino acid that helps the body get rid of isovaleric acid. It is often given as a supplement to children with IVA. It may help prevent metabolic crises. Your child’s metabolic specialist will prescribe this supplement and the correct amount to take, if necessary.
- Some children may benefit by taking L-carnitine. This is a safe and natural substance that helps the body make energy. It also helps the body get rid of isovaleric acid and other harmful wastes. The metabolic specialist will decide whether or not your child needs L-carnitine and, if so, the proper amount they should take.
- Do not make any changes to medications or diet without the approval and permission of the metabolic specialist and the dietician.
- Contact your child’s doctor immediately at the start of any illness. Children with IVA may need to be treated in a hospital to prevent serious health problems.

IF TREATED

With prompt and careful treatment, children with IVA have a good chance to live healthy lives with typical growth and development. However, even when treated, some children still have repeated occurrences of metabolic crisis which can lead to life-long learning problems or mental delays.